Giant Eccrine Spiradenoma of the Hand

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Since the work of Kersting and Helwig[1] in 1956, eccrine spiradenoma (ES) has been recognized as a benign sweat gland tumor. It most often occurs in the trunk and head-and-neck region but only very seldom in the hand. ES may present as a painful mass and may mimic other painful tumors, particularly angiolipomas. Incidence is highest in 15- to 35-year-olds.

ES usually occurs as a small (<1 cm) nodule with a bluish hue and less often as multiple discrete masses.[2-5] The differential diagnosis is often extensive, because of the nonspecific clinical and imaging findings. Other tumors that involve the upper extremity and may be confused with ES are lipoma, ganglion cyst, sebaceous cyst, giant cell tumor of tendon sheath, angiolipoma, and soft-tissue sarcoma. Although ES is usually benign, malignant transformation has been reported.[6-8]

We present the case of a patient with a benign ES involving the first dorsal web space of the left hand. Our patient was informed that data concerning his case would be submitted for publication.

Case Report

A 40-year-old right-hand–dominant Caucasian man presented with a long-standing history of a slowly enlarging painless mass on the dorsum of the left first web space. He first noticed the mass more than 20 years earlier, when his hand was struck with a heavy mallet. The 6×4-cm mass was in the superficial soft tissues (Figures 1A, 1B). There was a bluish discoloration on the skin, and the mass was slightly mobile. The mass was nontender to palpation, and there was no skin involvement. The left hand had no neurovascular deficits. The patient had no palpable lymphadenopathy of the antecubital or axillary lymph nodes and no other soft-tissue masses about the left upper extremity.

Radiographs showed a soft-tissue shadow without evidence of intralesional matrix or bone involvement (Figure 2). T_2-weighted magnetic resonance imaging (MRI) showed a mass with increased, heterogenous signal. T_1-weighted MRI showed that the mass was located superficial to the extensor tendons without invasion into adjacent neurovascular structures (Figure 3). A needle biopsy was performed under ultrasound guidance through a dorsal approach. Histology was interpreted as highly suspicious for a low-grade malignant spindle cell neoplasm, but additional tissue was requested for definitive diagnosis. An open biopsy was performed through a 1-cm dorsal approach, including the prior needle biopsy track. Microscopic evaluation showed a highly cellular tumor with epithelial cells arranged in intertwining bands. In addition, small cells with dark nuclei surrounded centrally located large cells with pale nuclei forming pseudoglandular rosettes (Figure 4). No malignant features (eg, mitotic figures, anaplasia) were present. The biopsy specimen was consistent with ES. Staging studies, including computed tomography of the chest, abdomen, and pelvis, showed no evidence of distant disease.

The patient underwent wide resection of the mass with a posterior interosseous artery flap for soft-tissue reconstruction (Figure 5). Additional options for soft-tissue coverage included split-thickness skin grafting alone or reverse-flow radial flap. The flap, selected according to anatomical location, provided sufficient soft-tissue coverage without significant donor site morbidity. The skin was outlined for a planned circumferential dissection, which included

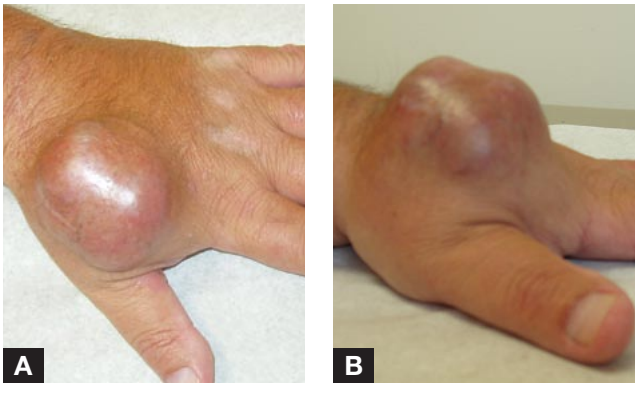

Figure 1. Dorsal (A) and profile (B) views of patient's left hand mass.

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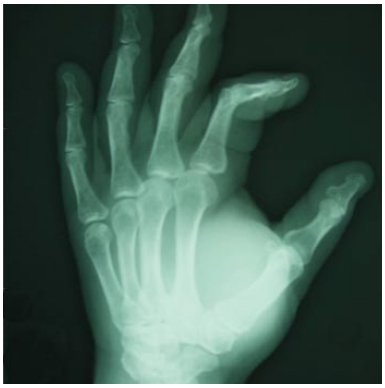

Figure 2. Oblique radiograph of left hand shows soft-tissue shadow in first web space without evidence of radiodense intralesional matrix.

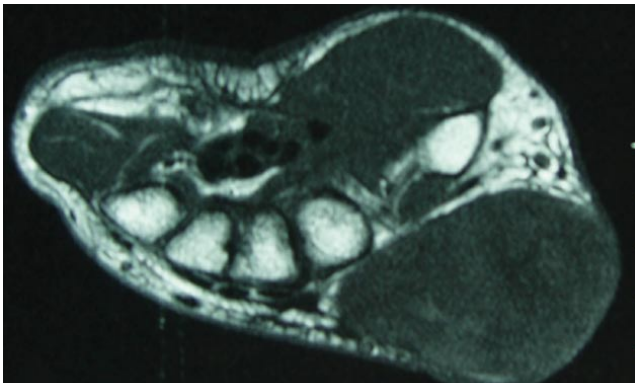

Figure 3. T_1-weighted axial image of left hand. The mass has low signal attenuation, and a plane of adipose tissue encompasses the deep surface.

an approximately 5-mm margin of the involved skin. The skin was incised circumferentially around the mass with sharp dissection through the subcutaneous tissue, with the intent to expose the underlying extensor pollicis longus. The radial and ulnar neurovascular bundles to the thumb were identified and carefully protected. The mass was mobilized away from the adjacent extensor pollicis longus and neurovascular bundle, preserving a 2- to 3-mm cuff of normal connective tissue circumferentially around the mass (Figure 5). On intraoperative frozen section sampling, the margins were found free of tumor. The defect was 6.5 cm (length) × 4.5 cm (width).

A separate setup, including drapes, gowns, and instrumentation, was used for the soft-tissue coverage aspect of the procedure. The posterior interosseous artery flap was started by identifying the intermuscular septum between the extensor digiti minimi and the extensor carpi ulnaris along with the septocutaneous perforators. A subcutaneous tunnel was then made from the distal incision point overlying the distal radioulnar joint above the extensor retinaculum and into the site of the defect. The flap was passed from proximal to distal, and at this point the flap was tacked into position around its periphery using interrupted sutures of 4-0 nylon.

After surgery, sensation to the radial and ulnar borders of the thumb was less than 5 mm to 2-point discrimination, and the thumb had full active abduction, opposition, flexion, and extension. Six weeks after surgery, the skin

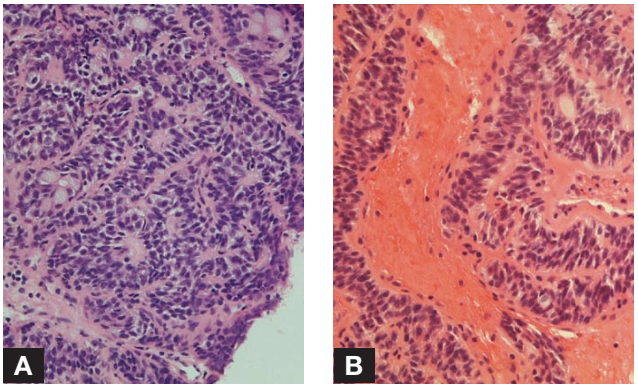

Figure 4. (A) Low-power photomicrograph shows basaloid tumor cells arranged in trabeculae and cords organized in ductlike configurations (hematoxylin-eosin, original magnification ×100). (B) Two types of cells are visible: larger, centrally located cells with pale cytoplasm and nuclei, with a tendency to aggregate and palisade around the lumina, and more peripherally located smaller cells with dark staining nuclei. Collections of clear cells were identified throughout the tumor (hematoxylin-eosin, original magnification ×100).

graft placed on the donor site was well healed and the flap completely viable.

Three years after surgery, the patient was back at his job and engaged in his usual activities (Figure 6). He had no evidence of local recurrence or antecubital/axillary lymphadenopathy. The most recent MRI scan of the hand showed no evidence of recurrence. The patient will continue to be followed annually with MRI plus contrast of the left hand.

Discussion

Eccrine sweat glands are simple tubular glands that open directly to the skin and are present in all body areas, with their highest concentration in the palms, soles, and axillae. Their thin, watery secretion is part of the heat-regulating mechanism of the body.[9] Etiology of ES is not well understood, and the very low incidence of this tumor may lead to a delay in diagnosis and treatment. Trauma has been implicated as an inciting factor, but no mechanism has been elucidated. Our patient had a distant history of trauma; whether it was coincidental or causative is unknown. Treatment requirements are not well understood, but recurrence (thought to result from inadequate resection) is rare.[2] ES may recur locally, produce deformity and pain, and in some cases transform into malignancies. Zamboni and colleagues[10] noted that ES involving the hand had a particularly high recurrence rate. Indicators of malignant transformation include rapid enlargement, bleeding, and ulceration.

Malignant ES, first described by Dabska in 1972,[11] can occur in Brooke-Spiegler syndrome. This syndrome manifests with cylindromas, ES, and trichoepitheliomas; its etiology is a defect in the CYLD gene on chromosome 9.[12] Malignant sweat gland tumors occur in the upper extremity. Metastasis may be lymphatic or hematogenous. The most common sites are lungs and regional lymph nodes, but bone, skin, brain, and kidneys have all been affected.[13]

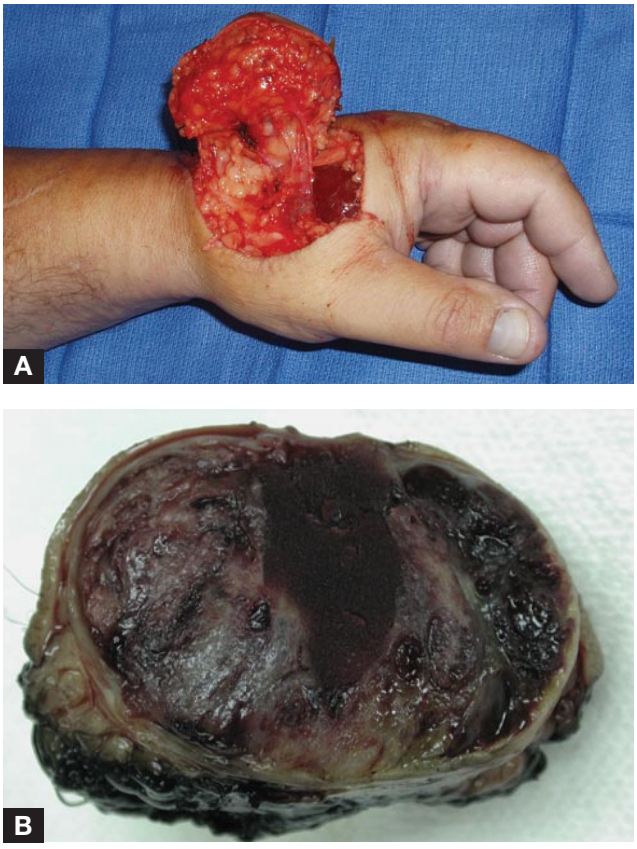

Figure 5. (A) The tumor is elevated away from underlying normal muscle and the neurovascular bundle. The tumor was removed with a covering of normal tissue. A large soft-tissue defect remained. (B) Cross-sectional view of gross specimen. A capsule is seen circumferentially around the mass. This yellowish-white capsule may give the false appearance of a lipoma.[15]

Our patient initially identified his mass more than 20 years before seeking treatment (he was asymptomatic during that period). Others investigators have suggested that long-standing disease may result in malignant transformation,[2,10,14] but this was not observed in our patient. Some patients present with solitary or multiple painful masses that may be confused with posttraumatic pain. In patients who do not recover from injury in a timely manner, further investigation with imaging and biopsy is indicated.

Ideally, the biopsy, whether needle or open, should be performed or supervised by the surgeon who, in case of malignancy, will be doing the definitive surgery. As in our patient's case, needle biopsy may not provide enough material for diagnosis. Malignant transformation has been reported to occur adjacent to benign ES, and, therefore, the potential for sampling error should be considered. Multiple core passes through the mass or open, incisional biopsy is recommended. If wide resection can be performed with minimal functional or cosmetic loss, it may be done without prior biopsy.

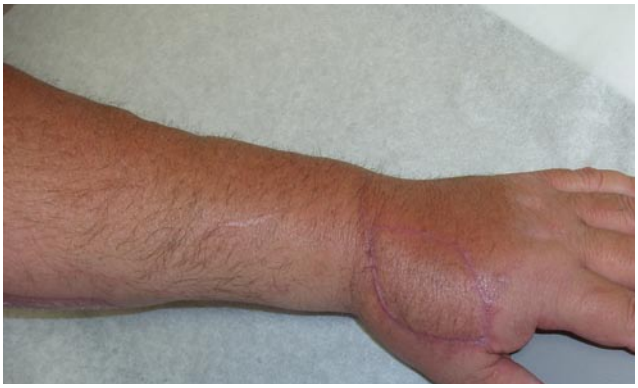

Figure 6. Two years after surgery, the patient had a well-healed soft-tissue flap and was again involved in his regular employment and activities, with no evidence of recurrence.

Although ES is benign, awareness of the entity is essential for timely diagnosis and treatment. Trauma history, pain, enlarged regional lymph nodes, and recent change in size or color should be noted. Close local surveillance (MRI plus contrast) for recurrence or malignant transformation is essential.[2,10,11]

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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